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## *Full Length* Case Report

# A CASE REPORT: NONSYNDROMIC FAMILIAL OLIGODONTIA

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**Abstract**

Oligodontia designates the congenital absence of six or more permanent teeth, excluding the third molars. The etiology behind this may be environmental and genetics factors. Genes responsible for non syndromicoligodontia are found to be MSX1 and PAX9 genes.The present case report describes a case of multiple agenesis of permanent teeth in a 14 year old female, with a positive family history. On examination, there was no abnormality in either hairs or nails, perspiration was normal and no congenital clefts of lip or palate was seen. Hence in this case it is not associated with any syndrome and was familial in nature.

**Keywords:** Familial, Oligodontia, Panoramic radiograph, Permanent Teeth



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## INTRODUCTION

Dental agenesis is the most common developmental anomaly in humans which can occur in an isolated fashion, or as a part of syndrome. Isolated cases of missing teeth can be familial or sporadic in nature (Ghazahfaruddin and Mishra, 2011). The incidence of such developmentally missing teeth might range from the absence of one or few teeth (hypodontia) to the absence of several teeth (oligodontia or severe hypodontia), or the failure of all teeth to develop (anadontia). The definition of oligodontia, which can occur as an isolated (nonsyndrome) condition or as a part of a syndrome, is the agenesis of six or more teeth, excluding the third molars and it is commonly seen in the permanent dentition rather than the deciduous dentition (Halicioglu *et al*., 2013). In previous studies, it was seen that more than 80% of hypodontia present with one or two congenitally missing teeth and only less than 1% will present with six or more. The incidence of oligodontia usually varies from 0.08% to 0.16% (Tangada and Batra, 2010) and prevalence of 0.3%[1]. It is a rare condition and the occurrence is common in girls in the ratio of 3:2. Oligodontia can occur in association with various genetic syndromes, like ectodermal dysplasia, Van Der Woude syndrome, Down syndrome and Reiger syndrome or as a nonsyndromic isolated familial trait. Syndromic and nonsyndromic form of oligodontia can be differentiated by conducting thorough physical examination of hair, nails, sweat glands, eyes, and checks for any congenital disorders. Treatment planning and space management for patients during the mixed dentition period must obviously include an evaluation of the number of teeth in both jaws. Knowledge of a congenitally absent tooth in one position should lead the clinician to consider the size and number of the remaining teeth (Dali *et al*., 2012). This case report presents a familial nonsyndromicoligodontia in a 14 yr old female patient with 15 permanent teeth missing.

## CASE REPORT

A 14 year old female reported to the department of pedodontics with a complaint of irregular teeth in upper front tooth region. Other than irregular teeth they also had a complaint of multiple missing teeth in both upper and lower arch. Her past medical history did not reveal any history of serious childhood illness or systemic abnormality and family history revealed that her father also had missing teeth in both the arches. No significant history was evident with her mother and other siblings (one younger sister and a brother). Intraoral examination revealed that there were many permanent teeth missing.The teeth clinically present were 16, 55, 54, 53, 11, 51, 21, 63, 64, 65, 26, 36, 75, 74, 73, 32, 71, 81, 42, 43, 44, 45, 56.The maxillary right central incisor was distally inclined, due to the retained deciduous right central incisor which showed preshedding mobility. The retained left and right canine and molars showed attrition with bilateral absence of permanent lateral incisors and canine. Normal maxillary right and left 1st permanent molars, with normal permanent left central incisor was evident. The mandibular arch revealed retained deciduous centrals which were severally attrited with the absence of permanent centrals.

There were erupting left premolars which were rotated and a stainless steel crown was present on right 2nd deciduous molar. Radiographically it was confirmed that 17, 15, 14, 13, 12, 22, 23, 24, 25, 27, 37, 35, 31, 41, 47 were missing (Figure 1). Similarly her father was examined to find out several teeth missing. The missing teeth which were evident with father were 13,15,25,31,32,35,36,37,41,42,43,44 and 45 (Figure 2). On examination of her mother and siblings revealed normal. Based on clinical and radiological examination, a diagnosis of familial nonsyndromicoligodontia was made. Retained 51 was extracted and the patient was referred to orthodontic department for closure of midline diastema was planned later for treatment partial denture for missing teeth.

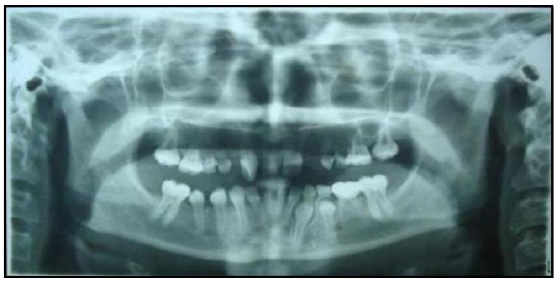

**Figure 1. Orthropantomograph showing missing teeth**

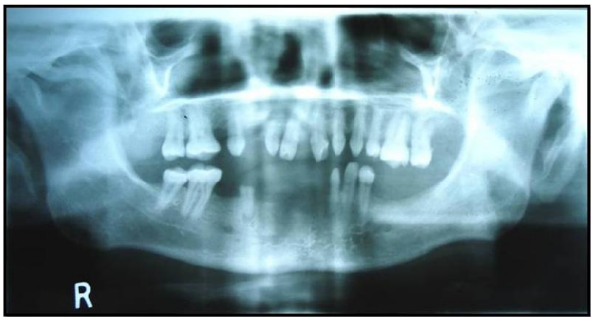


**Figure 2.**

## DISCUSSION

The etiology of dental agenesis is still not quite clear. Several hypotheses have been postulated (Polder *et al.,* 2004). Although environmental factors like infection, trauma, drugs, chemotherapy or radiotherapy have been considered as the etiology, but there is a strong evidence of genetic component. Oligodontia can be found as an isolated nonsyndromic trait or as a part of a syndrome, such as, ectodermal dysplasia, Down syndrome, incontinentiapigmentii and rieger syndrome. Mutation in the transcription factor MSX1 and PAX9 have been identified in families. The factors were demonstrated to be associated with isolated non-syndromicoligodontia. Recent studies have shown that mutation in EDA gene could result in non- syndromicoligodontia (Ghazahfaruddin *et al*., 2011). These isolated forms may be sporadic or familial. Familial tooth agenesis can be result of single dominant gene defect, a recessive or X-linked (Vastardis, 2000). Grahnen (Grahnen, 1956) has suggested that tooth agenesis is typically transmitted as an autosomal dominant trait with incomplete penetrance and variable expressivity. In his sample the penetrance was higher when the proband of the family had more than 6 missing teeth (Vastardis, 2000).

Brzynski and Escobar (Burzynski and Escobar, 1983) calculated the penetrance of numeric anomalies of dentition to be 86% with the use of Grahnen's data. Woolf (Woolf, 1971) has suggested that in families exhibiting dominant inheritance of incisor agenesis. This was also confirmed by Chosach *et al*. (1975) who reported that the incidence of missing lateral incisors was 9.7% in parents and 11.1% in siblings of index cases whose maxillary lateral incisors was also missing.

Other recent studies also supported that the maxillary lateral incisors are the most commolnly missing teeth in partial anodontia. Most hypodontic patterns are bilaterally symmetrical, except for the maxillary lateral incisor, where left tooth is more often missing than right. But in the present caseit is symmetrically missing (Graber, 2001). Variation of the number of congenitally absent teeth between maxilla and mandible also is open debate. Although majority of studies have reported more oligodontia in mandible, in contrary the present case showed more missing teeth in maxilla (Graber, 2001). Congenital agenesis of teeth can create dental and facial disfigurement, which can lead to social withdrawal, especially in adolescent years. Treatment in all the presented three cases includes multidisciplinary team approach of pedodontists, orthodontists, oral and maxillofacial surgeons, and prosthodontists to restore aesthetics, functional, and psychological reasons depending on the severity of the condition and patient's perceived need for care. Factors to be taken into account before treatment planning includes age of the patient, number and condition of retained teeth, number of missing teeth, condition of supporting tissues, the occlusion, and the interocclusal space (Dali *et al*., 2012 ). Endo *et al*. have concluded from their study that, before planning and implementing orthodontic treatment on a patient with congenital missing incisors, some factors such as retroclination of alveolar bone and reduced mandibular (Dali *et al*., 2012).

Oligodontia condition should not be neglected as it may result in various disturbances like abnormal occlusion, altered facial appearance which may cause psychological distress, difficulty in mastication and speech. Treatment depends on extent of hypodontia and should consist of interdisciplinary approach. Therefore early diagnosis is important in such conditions. Case oftooth agenesis should be recorded with complete clinical history including medical and radiological investigations to rule out any syndrome (Tangada and Batra, 2010).

### Conclusion

Patients of oligodontia should be carefully evaluated by the clinicians. Multidisciplinary treatment planning using established and emerging techniques should be considered. Also patients with tooth agenesis have to be emphasized not only the functional problems but also psychological distress, which require early diagnosis and timely management of oral health.

## REFERENCES

Burzynski N.J. and Escobar, V.H. 1983. Classification and genetics of numeric anomalies of dentition. *Birth Defects Orig Artic Ser.,* 19(1):95-106.

Chosack, A. and Eidelman, T. 1975. Hypodontia: a polygenic trait a family study among Israeli Jews. *J. Dent. Res.,* 54:16-9.

Dali, M., Singh, R. and Naulakha, D. Idiopathic nonsyndromic tooth agenesis:A report of rare three. Journal of Interdisciplinary Dentistry / Sep-Dec 2012 / Vol-2 / Issue-3

Ghazahfaruddin, M., Mishra, G., Haseebuddin, S. and Mishra, A. 2011. Oligodontia of permanent teeth: A Rare Case Report. *Indian J somatology*, 2(4):285-87.

Graber, 2001. orthodontics: Principle and practice. 3rd edition.

Grahnen H. 1956. Hypodontia in the permanent dentition: a clinical and geneticalinvestigation. *Odont Revy.,* 7:1-100.

Halicioglu, K., Sahin, H., Corekci, B., Irgin, C. and Toptas, O. 2013. Isolated oligodontia in monozygotic twins. *Europ J of Dent.,* 7(1):111-14.

Polder, B.J., Van't Hof, M.A., Van der Linden, F.P. and Kuijpers-Jagtman, A.M. 2004 .A meta-analysis of the prevalence of dental agenesis of permanent teeth. *Community Dent Oral Epidemiol,* Jun;32(3):217-26.

Tangada, P. and Batra, M. 2010. Non syndromicoligodontia: case report. Ethiop *J. Helth Sci.,* Nov; 22(3):219-21.

Vastardis H. 2000. The genetics of human tooth agenesis: New discoveries for understanding dental anomalies. *Am. J. Orthod Dentofacial. Orthop,* 117:650-6.

Woolf C. M. 1971 .Missing maxillary lateral incisors: a genetic study. *Am. J. Hum. Genet*., May;23(3):289-96.

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